Atlanto-axial tuberculosis: a report on five cases

RPB von Borrmann and RN Dunn
Department of Orthopaedics, University of Cape Town

Abstract

Introduction: High cervical tuberculosis is a rare condition, occurring in 0.3 to 1% of patients with tuberculous spondylitis. As a result, it is a condition that is often overlooked or misdiagnosed for a considerable period of time until a more obvious and potentially dangerous complication develops.

Aim: To review patients with atlanto-axial tuberculosis with regard to their presentation, management and outcome.

Method: Five cases of atlanto-axial tuberculosis were reviewed from their case notes and X-rays, and followed up for clinical review.

Results: The mean patient age was 30 (16-54) years. The mean time from presentation to diagnosis was 5.6 (3-12) months. All presented with neck pain and stiffness, with three patients ascribing the symptoms to previous trauma.

Two patients showed a neurological deficit on presentation. Three of the patients had tuberculosis of other sites. All patients had pre-vertebral swelling on plain lateral C-spine radiographs and a collection anterior to C1-2 on CT scan or MRI.

Three of these patients underwent surgery, one for anterior abscess drainage and diagnosis, one for stabilisation and one for non-contiguous spinal involvement. All received a minimum of six months of four-drug anti-tuberculous chemotherapy. One patient was HIV positive.

All patients did well with complete resolution of all neurology and instability, and good recovery of range of motion.

Conclusion: Despite being very rare, atlanto-axial tuberculosis carries the risk of instability and neurological compromise. A high index of suspicion is thus necessary, particularly in endemic areas, in patients presenting with neck pain.

Non-operative management with bracing and anti-tuberculous agents is adequate management, with surgery indicated for instability.

Introduction

Approximately 2 billion people world-wide have tuberculosis. It is endemic in developing countries, and the AIDS pandemic has resulted in its re-emergence in developed countries as well.

Pulmonary TB is the most common presentation but spinal TB is the most common extrapulmonary presentation. According to Miller and Brinker 5 to 15% of osseous involvement is in the spine. Tuberculous spondylitis is found in less than 1% of patients with TB and atlanto-axial TB (AATB) has been diagnosed in only between 0.3 and 1% of all tuberculous spondylitis presentations, making this an extremely rare condition.

AATB causes extensive osteo-ligamentous destruction of the most mobile segment of the cervical spine. Neural compression, which often results, can be the result of either instability or direct compression by mass effect.

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AATB is often only recognised when complications, ranging from minor neurological deficits to sudden death, develop. The sum total of this is a very rare disease entity, and accordingly a diagnosis which is often overlooked resulting in considerable delays between presentation and diagnosis.

In this study we present five cases of AATB.

**Patients and methods**

Five patients were managed between July 2001 and October 2004. All were adults with a mean age of 30 years (16-54).

Patients were reviewed using their case notes, radiological studies and follow-up clinical assessment for a mean of 13.4 months (7-18).

**Presenting symptoms**

All the patients presented with neck pain and stiffness, two mistakenly attributing this to recent trauma. One had torticollis.

Only three of these patients complained of systemic symptoms. Three had weight loss and two reported having night sweats.

**Clinical findings**

Examination revealed generalised lymphadenopathy in three patients. All had demonstrable neck stiffness and tenderness posteriorly over the upper C-spine. In two patients there was a palpable neck mass. One was palpable posteriorly, and the other had a retro-pharyngeal mass causing dysphagia and respiratory compromise.

Only two patients had any neurological deficit. One had 4/5 weakness in the left arm with altered sensation and brisk reflexes. The other patient had bilateral upper and lower limb 4/5 weakness with increased tone and reflexes.

**Laboratory investigations**

We found white-cell counts unhelpful. The Erythrocyte Sedimentation Rate was normal in two cases of which one was a partially treated case and the other a retroviral positive patient.

**Radiological findings**

On plain X-rays all five cases demonstrated anterior soft tissue swelling. Three cases showed evidence of bony destruction involving the dens and body of C2 with a concomitant increase in the atlanto-dens interval.

All the patients had MRI scans, which showed altered bony signals with variable bony destruction and an abscess anterior to the atlanto-axial area.

Three of the five patients had TB of other sites demonstrated both clinically and radiologically. One patient had non-contiguous spinal involvement in the lower cervical spine and upper thoracic spine. One patient had chondrolysis of the left hip. The third had xiphisternal tuberculous osteitis.

**Diagnosis**

All five cases had a significant delay in diagnosis with a resultant delay in referral to our centre. The mean delay was 5.6 months (3-12).

One of these patients was incorrectly diagnosed as having a traumatic lesion, and treated in a POP jacket for six months by a peripheral hospital without chemotherapy.

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**Figure 1**

Case 1: This MRI demonstrates the classic anterior soft tissue abscess of atlanto-axial tuberculosis. This patient was successfully treated conservatively.

**Figure 2**

Case 2: This X-ray demonstrates bony destruction involving the dens and the body of C2 with resultant instability.
We achieved a positive TB diagnosis in all our cases following a biopsy. Four of the samples were positive for acid-fast bacilli on microscopy and one was confirmed on histology.

**Management**

All patients received four-drug anti-TB chemotherapy for a minimum of six months or until clinical and radiological union had been achieved (rifampicin 10-20 mg/kg, isoniazid 5-10 mg/kg, pyrazinamide 20-35 mg/kg, ethambutol 15 mg/kg).

All patients were immobilised in a brace. We used Halo, SOMI and Philadelphia braces.

Three patients had the soft tissue abscess anterior to C1-2 drained surgically. One of these was done via a transoral approach by the ENT department prior to referral to the spinal unit.

The other two were done by following the Smith-Robinson approach (anterior-lateral) by the senior author (RN Dunn). This was done for both therapeutic and diagnostic purposes.

In addition, the patient with the non-contiguous C-spine involvement underwent an anterior decompression and instrumented fusion of C4-7. No attempt was made to drain the abscess anterior to C1-2.

One patient underwent a posterior on-lay bone graft and fusion of occiput-C3 and halo-jacket immobilisation after an anterior biopsy. This was done because of severe bony destruction and instability both at occiput-C1 and at C1-2. She went on to solid occipito-cervical spine fusion.

**Outcomes**

All five patients went on to full neurological recovery. All achieved radiological union and were also stable on dynamic radiology.

Clinically cervical range-of-movement showed varied degrees of recovery, from normal to 50% of normal rotation. The patient with an atlanto-axial rotatory subluxation remained with a fixed torticollis.

**Discussion**

**Presentation and clinical findings**

Our patient presentation was in keeping with reports in the literature as regards delays in presentation, presenting complaints and clinical findings. In contrast to Behari et al, who found that 72% of their patients had neurology at presentation, we found only 40% in our group.

**Imaging**

Behari et al. found that X-ray changes in tuberculous spondylitis lagged behind the pathological alterations in architecture by from two to six months because bone erosion is only evident radiologically after 50% of the vertebra has been destroyed. They recommended the use of CT and MRI of the cranio-cervical junction in early atlanto-axial tuberculosis for both diagnosis and planning as this demonstrated bony fragmentation at the vertebral end plates and surrounding soft tissue changes. They also described the finding of a multilocular, calcified abscess with a thick, enhancing, irregular rim in the presence of vertebral body fragmentation as pathognomonic of TB. De Vuyst et al. describe similar MRI findings and put forward that MRI is the imaging modality of choice for diagnosis.

All our cases had anterior soft tissue swelling changes on initial lateral C-spine X-rays. This however is possibly explained by the delay in presentation. The MRI changes of our patients were in keeping with Behari’s findings.

**Diagnosis**

Crockard et al. emphasise the importance of tissue diagnosis to reliably distinguish TB from malignancy and to prevent multi-drug resistance.
Management

Management has swung between absolute conservatism and radical surgery. Earlier authors were more aggressive.\(^\text{6,7,8,9}\) Fang\(^\text{10}\) reported six cases of atlanto-axial tuberculosis for which he was in favour of a surgically aggressive regimen, recommending a transoral decompression and anterior fusion. Despite indicating that conservative management is hazardous, a further twelve cases had been managed without surgery. He did not report on their outcome.

Lifesø et al\(^\text{11}\) offered a classification based on bony destruction and instability. A transoral biopsy was recommended in all, but he reserved fusion surgery for cases with anterior displacement of C1 on C2 or significant bony destruction. For this he used a transoral decompression and posterior fusion, extending to the occiput if there was marked bony destruction or instability.

Surgical approach

The literature is almost unanimously in favour of decompressing the soft tissue abscess through a transoral approach.\(^\text{5,7,8,11}\)

Recommendations regarding the approach to fusion vary. The transoral approach was advocated by most authors, including Lifeso\(^\text{9,10}\) and is still regarded as the approach of choice by Crockard et al\(^\text{12}\) and others.\(^\text{6,7,11}\) They maintain that this approach provides excellent access to this region with low operative morbidity and mortality.

Concerns about bacterial contamination and secondary pyogenic infection did, however, lead Sinha et al\(^\text{13}\) to prescribe a transcervical retropharyngeal approach for abscess decompression. This paper on 18 patients and his experience of 15 years shows that the transcervical retropharyngeal approach gives wide exposure and direct access to occiput-C2. Bacterial contamination of the oral and pharyngeal cavities is avoided. He also notes that the transoral approach can lead to persistent sinus formation.

Tuli\(^\text{1}\) also shows that the transcervical Smith-Robinson approach is adequate for abscess evacuation. This was also borne out in our experience.

Current trends

There is a swing back to more conservative management as emphasised by Behari et al,\(^\text{1}\) which is the most comprehensive study of atlanto-axial tuberculosis, namely comprising 25 patients. These authors showed that often the only treatment needed is anti-tuberculous chemotherapy and immobilisation. The granulation tissue resolves and the destroyed bone heals.\(^\text{14}\) They reserve anterior decompression and posterior stabilisation for severe myelopathy.

Summary

Despite being very rare, atlanto-axial tuberculosis carries the risk of instability and neurological compromise. A high index of suspicion is thus necessary, particularly in endemic areas, when patients present with neck pain.

In our experience conservative management yielded full neurological recovery, atlanto-axial stability and resolution of the soft tissue abscess.

For both biopsy, and when necessary, abscess evacuation, we used the anterior cervical retropharyngeal Smith-Robinson approach.

We recommend that surgical stabilisation be reserved for severe osseo-ligamentous destruction and instability.

References